Seborrheic dermatitis was also common (n = 43, 21.8%) (Table II).

Other comorbidities included Vitamin D deficiency (n = 43, 21.8%), hyperlipidemia (n = 39, 19.8%), obesity (n = 35, 17.8%), gastroesophageal reflux disease (n = 32, 16.2%), and anemia (n = 24, 12.2%). Psychiatric comorbidities including depression, anxiety, or sleep disorders were identified in 14.2%, with absence of documented history of suicide attempts (Table II).

Our study shows that H/L patients with AA have a similar average age at diagnosis, slight female predominance, and increased atopy compared to the general population in recent AA studies.^{1,2} Male H/L patients with AA were younger than female at diagnosis (P = .01, Table I). Interestingly, the most common autoimmune comorbidity in H/L was rheumatoid arthritis, compared to thyroid disease in Black patients¹ and overall patients with AA.^{2,3} This finding may be a reflection of a larger trend, as rheumatoid arthritis in the H/L population has been on the rise.⁴ Thyroid disease in our H/L cohort with AA was slightly higher than the 4.8% reported in the general Hispanic population.⁵ Atopy and hypertension were less common in H/L than in Black patients¹ or patients with AA overall, but gastroesophageal reflux disease was more prevalent.³

A strength of our study is that most patients were evaluated by a hair specialist on the study team (NAM). Limitations include the small sample size, lack of a control group, and that patients may have received additional healthcare outside of our center. The study findings increase the current knowledge of the demographics of H/L patients with AA, and heighten awareness of associated inflammatory comorbidities, in particular rheumatoid arthritis.

Celine H. Phong, BS,^a Amy J. McMichael, MD,^b and Natasha Atanaskova Mesinkovska, MD, PhD^a

From the Department of Dermatology, University of California, Irvine, Irvine, California^a; and Department of Dermatology, Wake Forest School of Medicine, Winston-Salem, North Carolina.^b

Funding sources: None.

IRB approval status: Reviewed and approved by the University of California, Irvine (2015-2022) IRB (IRB 2016-3076).

Patient consent forms: Not applicable.

Key words: alopecia areata; alopecia totalis; alopecia universalis; comorbidities; epidemiology; Hispanic; Latina; Latino; Latinx.

Correspondence and reprint requests to: Natasha Atanaskova Mesinkovska, MD, PhD, Department of Dermatology, University of California, Irvine, Hewitt Hall, 843 Health Sciences Rd, Room 1001, Irvine, CA 92697

E-mail: natashadermatology@gmail.com

Conflicts of interest

None disclosed.

REFERENCES

- 1. Feaster B, McMichael AJ. Epidemiology of alopecia areata in Black patients: a retrospective chart review. J Am Acad Dermatol. 2022;87(5):1121-1123. https://doi.org/10.1016/j.jaad. 2022.01.033
- Conic RZ, Miller R, Piliang M, Bergfeld W, Atanaskova Mesinkovska N. Comorbidities in patients with alopecia areata. *J Am Acad Dermatol.* 2017;76(4):755-757. https://doi.org/10. 1016/j.jaad.2016.12.007
- Huang KP, Mullangi S, Guo Y, Qureshi AA. Autoimmune, atopic, and mental health comorbid conditions associated with alopecia areata in the United States. *JAMA Dermatol.* 2013;149(7):789. https://doi.org/10.1001/jamadermatol.2013. 3049
- Xu Y, Wu Q. Prevalence trend and disparities in rheumatoid arthritis among US adults, 2005–2018. J Clin Med. 2021;10(15): 3289. https://doi.org/10.3390/jcm10153289
- Hollowell JG, Staehling NW, Flanders WD, et al. Serum TSH, T4, and thyroid antibodies in the United States population (1988 to 1994): National Health and Nutrition Examination Survey (NHANES III). J Clin Endocrinol Metab. 2002;87(2):489-499. https://doi.org/10.1210/jcem.87.2.8182

https://doi.org/10.1016/j.jaad.2022.10.049

Clinical and pathological features of cutaneous manifestations in VEXAS syndrome: A multicenter retrospective study of 59 cases



To the Editor: VEXAS (vacuoles, enzyme E1, X-linked, auto-inflammatory, somatic) syndrome is a late-onset autoinflammatory condition due to myeloid-restricted somatic mutations in the ubiquitin-activating enzyme 1 gene.¹ Skin involvement seems to be one of the most common symptoms, ¹⁻⁴ and may be the first manifestation of VEXAS syndrome.⁵ However, most studies have been performed in non-dermatology departments with no centralized review of cutaneous involvement and limited numbers of patients.^{4,5}

In this multicenter nationwide retrospective study, all 59 patients from the French VEXAS study group (NFVS) database with photographs of skin lesions and/or skin biopsies available for centralized review on June 15, 2021, were included. This study received approval from our Institutional Review Board (CLEP Decision no.:AAA-2021-08040). -----

$\operatorname{ment}(n=57)$	
Lesion description	
Lesion type	
Maculopapules and nodules	37 (100%)
Pustules	5 (13%)
Vesicles/bullae	4 (11%)
Livedo	6 (16%)
Reticularis	3 (8%)
Racemosa	3 (8%)
Thickness of pattern <1 cm	3 (8%)
Lesion shape	
Round/nummular	36 (97%)
Arcuate/annular	12 (32%)
Lesion color	
Pink	28 (76%)
Red	27 (73%)
Violaceous/purpuric	18 (49%)
Lesion size	
<1 cm	29 (78%)
≥1 cm	29 (78%)
Both	21 (57%)
Number of lesions	
≥10	33 (89%)
<10	4 (11%)
Localization	
Trunk	30 (81%)
Arms	32 (86%)
Legs	31 (84%)
Face	11 (30%)
Pathergy	5 (13%)
Cutaneous symptoms	
Pain	13 (35%)
Pruritus	9 (24%)
Evolution of skin lesions	
Flare/remission periods	30 (81%)
Frequency of flare-ups	6 (2-12)
(times/y, median [IQR])	
Duration of flare-ups (d, median [IQR])	9.8 (7-10)
Permanent	4 (11%)

Table I. Clinical characteristics of skin involve-

IQR, Interquartile range.

The median age at first symptoms was 68 years (interquartile range: 61.5-73) and 97% patients were male (Supplementary Table I, available via Mendeley at https://doi.org/10.17632/zpz5h43c4w.1). Cutaneous involvement was the first symptom for most patients (63%). The most common initial hypothesis for the skin involvement was Sweet syndrome (47%). Among the 37 patients with available photographs (Table I), all showed round shaped maculopapules of various sizes (57% showed both < and \geq 1-cm lesions), mostly pink/ red (76%), often numerous (89% of patients had \geq 10 lesions), on the trunk (81%), the limbs (85%), and the face (30%). Lesions were arcuate in approximately one-third (32%, Fig 1). Other cutaneous lesions included



Fig 1. Arcuate papules in VEXAS (vacuoles, enzyme E1, X-linked, auto-inflammatory, somatic) syndrome.

livedo (16%), pustules (13%), and injection-site reactions (pathergy, 13%) (Supplementary Fig 1, available via Mendeley at https://doi.org/10.17632/ zpz5h43c4w.1). Skin lesions were typically characterized by periods of flare and remission (81%). Among the 43 patients with available biopsies, most (89%) patients had infiltrates of the superficial dermis. The infiltrates consisted of immature myeloid cells and variable proportions of mature neutrophils, lymphocytes, and histiocytes, which led to retaining the diagnosis of neutrophilic dermatosis rich in immature myeloid cells in 77% patients (Supplementary Table II, available via Mendeley at https://doi.org/10.17632/ zpz5h43c4w.1). Leukocytoclasia was present in 63% patients; however, no vasculitis was found. Few patients showed vessel thrombosis (venous in 3 patients, capillary in 2 patients, and arteriolar in 1 patient). Immunostaining revealed the presence myeloid precursors with overlapping expression of CD68 and MPO (myeloperoxidase) in 88% (Supplementary Fig 2, available via Mendeley at https://doi.org/10.17632/zpz5h43c4w.1). Cytological analysis was performed in 30 cases to better characterize the myeloid precursors. They were classified as metamyelocytes in 52%, as myelocytes in 42%, and as band neutrophils in 26%.

Overall, the spectrum of cutaneous lesions of VEXAS is characterized by a wide heterogeneity. About one-third of patients showed lesions with an arcuate shape (Fig 1), which we believe is suggestive of the diagnosis. Pathological infiltrates typically consisted of superficial perivascular and periadnexal lymphohistiocytic infiltrates with immature myeloid cells and displayed signs of dysplasia in most cases. In previous studies, vasculitis was frequently reported (25%-70%),^{1,3-5} but this rate has probably been overestimated. Indeed, despite the high prevalence of leukocytoclasia, we found no vasculitis after centralized review. The main limitation of our study is its retrospective design.

In conclusion, VEXAS syndrome should be considered in patients ≥ 60 years with general symptoms, systemic inflammation, and with skin infiltrates showing neutrophilic dermatosis with immature myeloid cells.

We thank the French National FAI2R network for their help in realizing this study, and the physicians involved in the French Vasculitis Study Group, in CeReMAIA and MINHEMON. We thank all the contributors that made this study possible by providing clinical photographs and organizing pathology material dispatch: Z. Amoura (Pitié-Salpêtrière), A. Aouba (Caen), C. Arnaud (Toulouse), A. Audemard-Verger (Tours), C. Bachmeyer (Tenon), B. Bienvenu (Marseille), P. Biscay (Pessac), F. Borlot (Béziers), L. Bouillet (Grenoble), G. Boursier (Montpellier), F. Carrat (Saint Antoine), T. Cluzeau (Nice), T. Comont (Toulouse), A. Constantin (Toulouse), B. de Sainte Marie (Bordeaux), C. Deligny (Martinique), C. Dieval (Rochefort), E. Duroyon (Centre), M. Ebbo (La Timone), O. Fain (Saint Antoine), B. Faucher (La Timone), P. Fenaux (Saint Louis), S. Georgin-Lavialle (Tenon), M. Gerfaud-Valentin (Hospices Civils de Lyon), J. Graveleau (Nantes), A.F. Guedon (Saint Antoine), T. Hanslik, (Ambroise Paré), M. Heiblig (Hospices Civils de Lyon), V. Jachiet (Saint Antoine), Y. Jamilloux (Hospices Civils de Lyon), J. Jeannel (Paris), M. Larue (Mondor), F. Le Pelletier (Paris), E. Liozon (Limoges), A. Meyer (Strasbourg), T. Moulinet (Nancy), M. Pha (Pitié-Salpêtrière), J. Rossignol (Necker), M. Roux (Nord Dauphiné), M. Roux-Sauvat (GHND), L. Sailler (Toulouse), G. Sarrabay (Montepellier), M. Sebert (Saint Louis), A. Servettaz (Reims), P. Sujobert (Besançon), L. Terriou (Lille), J. Vinit (Chalons), S. Vinzio (Grenoble), T. Weitten (Gap), L.P. Zhao (Saint Louis). We thank Laura Smales English language editing.

 Ève Zakine, MD,^a Loula Papageorgiou, MD,^b Rim Bourguiba, MD,^c Arsène Mekinian, MD, PhD,^d Benjamin Terrier, MD, PhD,^e Olivier Kosmider, MD, PhD,^f Pierre Hirsch, MD,^b Marie Jachiet, MD,^a Sylvain Audia, MD, PhD,^g Samuel Ardois, MD,^b Léopold Adélaïde, MD,ⁱ Adrien Bigot, MD,^j Paul Duriez, MD,^k Jean-François Emile, MD, PhD,¹ Estibaliz Lazaro, MD, PhD,^m Damien Fayard, MD,ⁿ Joris Galland, MD,^o Miguel Hié, MD,^p Sébastien Humbert, MD,^q Alexis Jean, MD,^m Marie Kostine, MD,^r Valentin Lacombe, MD,^s Guillaume Le Guenno, MD,^t Hervé Lobbes, MD,^t Nadine Magy-Bertrand, MD, PhD,^q Paola Marianetti-Guingel, MD,^u Alexis Mathian, MD,^p Rodérau Outb, MD,^v Clémence Saillard, MD,^w Maxime Samson, MD, PhD,^g Guillaume Vial, MD,^m Jean-David Bouaziz, MD, PhD,^a Philippe Moguelet, MD,^x and François Chasset MD, PhD,^y National French VEXAS Study Group (NFVS)

From the Dermatology Department, Paris University, Faculty of Medicine, AP-HP, Saint Louis University Hospital, Paris, France^a; Hematology and Cytology Department, Sorbonne University, INSERM, Saint Antoine Research Center, AP-HP, Saint Antoine University Hospital, Paris, France^b; Internal Medicine Department, Sorbonne University, Faculty of Medicine, AP-HP, Tenon University Hospital, Paris, France^c; Internal Medicine Department, Sorbonne University, Faculty of Medicine, AP-HP, Saint Antoine University Hospital, Paris, France^d; Internal Medicine Department, Paris University, Faculty of Medicine, AP-HP, Cochin University Hospital, Paris, France^e; Hematology Department, Paris University, Faculty of Medicine, AP-HP, Cochin University Hospital, Paris, France^f; Internal Medicine Department, Bourgogne University, Faculty of Medicine, Dijon University Hospital, Dijon, France^g; Internal Medicine Department, Rennes University, Faculty of Medicine, Rennes University Hospital, Rennes, France^b; Internal Medicine Department, Claude Bernard Lyon 1 University, Faculty of Medicine, Croix-Rousse Hospital, Lyon, France^{i;} Internal Medicine Department, Tours University, Faculty of Medicine, Tours University Hospital, Tours, France^{*i*}; Pathology Department, Sorbonne University, Faculty of Medicine, AP-HP, Saint Antoine University Hospital, Paris, France^k; Pathology Department, Versailles Saint Quentin University, Faculty of Medicine, Ambroise Paré Hospital, Boulogne-Billancourt, France¹; Internal Medicine Department, Bordeaux University, Faculty of Medicine, Bordeaux University Hospital, Bordeaux, France^m; Internal Medicine Department, Clermont-Ferrand University, Faculty of Medicine, Gabriel Montpied University Hospital, Clermont-Ferrand, Franceⁿ; Internal Medicine Department, Fleyriat Hospital, Bourg-en-Bresse, France^o; Internal Medicine Department, Sorbonne University, Faculty of Medicine, AP-HP, Pitié-Salpêtrière

University Hospital, Paris, France^p; Internal Medicine Department, Besançon University, Faculty of Medicine, Besançon University Hospital, Paris, France^q; Rheumatology Department, Bordeaux University, Faculty of Medicine, Hospital, Bordeaux University Bordeaux, France^r; Internal Medicine Department, Clermont-Ferrand University, Faculty of Medicine, Estaing University Hospital, Clermont-Ferrand, France^s; Internal Medicine and Clinical Immunology Department, Angers University, Faculty of Medicine, Angers University Hospital, Angers, France^t; Internal Medicine Department, Reims University, Faculty of Medicine, Reims University Hospital, Reims, France^u; Internal Medicine Department, Perpignan Hospital Center, Perpignan, France^v; Dermatology Department, Rennes University, Faculty of Medicine, Rennes University Hospital, Rennes, France^w; Pathology Department, Sorbonne University, Faculty of Medicine, AP-HP, Tenon University Hospital, Paris, France^x; and Dermatology and Allergology Department, Sorbonne University, Faculty of Medicine, AP-HP, Tenon University Hospital, Paris, France.^y

Drs Moguelet and Chasset contributed equally and share senior authorship.

Funding sources: None.

- IRB approval status: Institutional Review Board (CLEP Decision no.:AAA-2021-08,040).
- Key words: dermatopathology; neutrophilic dermatoses; Sweet's syndrome; VEXAS syndrome.
- Reprint requests: François Chasset, MD, PhD, Service de Dermatologie et d'Allergologie, AP-HP, Sorbonne Université, Hôpital Tenon, 4 Rue de la Chine, 75970 CEDEX 20 Paris, France

E-mail: francois.chasset@aphp.fr

Conflicts of interest

None disclosed.

REFERENCES

- Beck DB, Ferrada MA, Sikora KA, et al. Somatic mutations in UBA1 and severe adult-onset autoinflammatory disease. N Engl J Med. 2020;383:2628-2638.
- 2. Georgin-Lavialle S, Terrier B, Guedon AF, et al. Further characterization of clinical and laboratory features in VEXAS syndrome: large-scale analysis of a multicentre case series of 116 French patients. *Br J Dermatol.* 2021;186(3):564-574. https://doi.org/10.1111/bjd.20805
- **3.** van der Made CI, Potjewijd J, Hoogstins A, et al. Adult-onset autoinflammation caused by somatic mutations in UBA1: a Dutch case series of patients with VEXAS. *J Allergy Clin Immunol*. 2022;149(1):432-439.e4.

- Sterling D, Duncan ME, Philippidou M, Salisbury JR, Kulasekararaj AG, Basu TN. VEXAS syndrome (vacuoles, E1 enzyme, X-linked, autoinflammatory, somatic) for the dermatologist. J Am Acad Dermatol. 2022. https://doi.org/10.1016/ j.jaad.2022.01.042
- Zakine E, Schell B, Battistella M, et al. UBA1 variations in neutrophilic dermatosis skin lesions of patients with VEXAS syndrome. JAMA Dermatol. 2021;157(11):1349-1354.

https://doi.org/10.1016/j.jaad.2022.10.052

High compliance with National Comprehensive Cancer Network guidelines and no local recurrences for patients receiving Mohs micrographic surgery for Merkel cell carcinoma: A single-center retrospective case series

To the Editor: National Comprehensive Cancer Network (NCCN) guidelines for Merkel cell carcinoma (MCC) recommend excision of the primary tumor and sentinel lymph node biopsy (SLNB) for staging. NCCN guidelines include Mohs micrographic surgery (MMS) as an excision technique and recommend sending a debulking excision for staging with paraffin-embedded vertical permanent sections.¹

Table I. Patient demographics and case characteristics for patients with Merkel cell carcinoma treated with Mohs micrographic surgery (n = 30)

Mean age, y (SD)	72.3 (16.0)
Female sex, n (%)	16 (53.3)
Immunosuppressed, n (%)	2 (6.7)
Tumor location, n (%)	
Cheek	8 (26.7)
Nose	7 (23.3)
Periorbital	5 (16.7)
Forehead	3 (10.0)
Ear	3 (10.0)
Extremities (excluding hands/feet)	2 (6.7)
Temple	2 (6.7)
Primary lesion, n (%)	26 (86.7)
Pre-op lesion size in cm, n (%)	
0.00-0.49	4 (13.3)
0.50-1.00	10 (33.3)
1.01-2.00	10 (33.3)
>2.00	6 (20.0)
Post-op lesion size in cm, n (%)	
0.50-1.00	1 (3.3)
1.01-2.00	6 (20.0)
2.01-5.00	18 (60.0)
5.01-10.00	5 (16.7)

post-op; Postoperative; *pre-op*, preoperative; *SD*, standard deviation.